Published Quarterly.

Part XCVII. [*Price 3s. 6d.*

BRAIN:

A JOURNAL OF NEUROLOGY

EDITED

FOR THE NEUROLOGICAL SOCIETY OF LONDON

BY

R. PERCY SMITH, M.D.

Assisted in the selection of Papers by

THOMAS BUZZARD, M.D.
VICTOR HORSLEY, M.B., F.R.S.
W. JULIUS MICKLE, M.D.
F. W. MOTT, M.D., F.R.S.
J. S. RISIEN RUSSELL, M.D.
GEORGE H. SAVAGE, M.D.
ERNEST H. STARLING, M.D., F.R.S.
AUGUSTUS WALLER, M.D., F.R.S.

NOTICE.—All Editorial Communications, Manuscripts, and Printed Matter may be addressed to the EDITOR, 36, Queen Anne Street, W.

London:
MACMILLAN AND CO., LTD.
NEW YORK: THE MACMILLAN CO.
SPRING, 1902.

INTERNAL HYDROCEPHALUS IN THE ADULT, WITH REMARKS ON THE ÆTIOLOGY OF HYDROCEPHALUS AND ITS OCCASIONAL ASSOCIATION WITH OTHER ABNORMAL CONDITIONS OF THE CENTRAL NERVOUS SYSTEM.

BY F. PARKES WEBER, M.D., F.R.C.P., LONDON,
Physician to the German Hospital.

A THIN and rather delicate-looking young woman, B.S., aged 22, single, a native of Saxony, was admitted into the German Hospital, June 10, 1901. She had already been ill about nine or ten weeks, suffering from headache, giddiness, and recurrent vomiting, and had been treated for gastric disorder.

Past History.—She was always healthy as a child. She was not subject to headache, and although she seldom took alcoholic stimulants, there was no evidence that alcohol affected her in any unusual way. No history of temporary deafness. She had twice suffered from erysipelas of the head. The first time was in her seventeenth year, and after the illness for a time she lost all the hair of her head. The second attack was a few years ago. During this she suffered from headache, and was kept three months in a hospital in Hanover. Afterwards she seemed well again. It is worth mentioning, however, that in the spring of 1901, when crossing from Hamburg to London, with the sea-sickness she seems to have lost, or almost lost, consciousness, being in a condition perhaps resembling the rare swooning form of *mal de mer*, and possibly suggesting the presence of some hitherto latent disease.

In the German Hospital examination of the gastric contents was reported to show absence of free hydrochloric acid and presence of lactic acid. When, however, the patient had been some time under observation the character of the vomiting was found to suggest a nervous origin. An attack would come on suddenly without accompanying gastric discomfort, especially after sudden movements. By ophthalmoscopic examination (July 15, 1901) distinct but slight optic neuritis was found in

both eyes (on the right side more than the left). Stiffness in the cervical region of the spine was complained of soon after admission, at one time suggesting caries in the upper cervical vertebræ. The stiffness was more marked for rotation than for flexion of the head. There was no evidence of disease in the thoracic or abdominal organs, excepting the gastric disorder noted above. The patient did not seem to be hysterical. She was decidedly intelligent and quick at understanding, and could sing. There was no evidence of syphilis, either congenital or acquired. With the exception once or twice of an evening temperature of 100°F. fever was absent throughout.

Mercurial inunction was tried with doubtful result. The patient, however, gradually improved; the cervical rigidity completely disappeared, and she was able to be up and get about in the ward and was thinking of leaving the hospital.

In November the patient's troubles to some extent returned, and mercurial inunction was tried again. Pepsin was likewise given to aid digestion. Dr. Leonard Williams, who kindly took charge of the case during my absence, found her complaining of headache, vomiting and weakness. There was slight nystagmus on extreme lateral deviation, which was ill sustained. Optic neuritis in both eyes. The knee jerks were present, equal, and active. No clonus. The plantar reflex was of the flexor type. Dr. Williams found slight ataxia in one of the upper limbs. Vertigo was not a very prominent symptom of the case, but the sister of the ward says the patient on lying down after a bath would sometimes feel giddy and then suddenly vomit. Her gait was hesitating, rather that of a weak person taking small steps than that of a patient with cerebellar tumour or tabes. Deafness was not a symptom in the case.[1] Neither was there any mental disturbance such as occurs in general paralysis, nor any form of insanity such as has been observed in some cases.[2] On December 7, 1901, at about 6.30 a.m., the patient was suddenly seized with violent vomiting and frightful headache, and this was followed by general rigidity and clonic convulsive movements, death occurring about 7.30 a.m. The pulse during the seizure was infrequent but regular, and the face was flushed at the end.

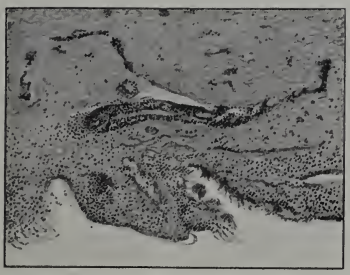
[1] As it was in Hilton's case and in that of Burr and McCarthy, both of which I have quoted later on.

[2] See Burr and McCarthy's case quoted later on. See also Dr. T. Whipham's account of a man, aged 39, with hydrocephalus and insanity (*Trans. Clinical Soc. of London*, 1884, vol 17, p. 191.) Dr. Whipham's case was not merely one of passive hydrocephalus or "hydrocephalus ex vacuo," as there was flattening of convolutions over a portion of the brain.

At the *post-mortem examination* the cerebral convolutions were found much flattened, and all the ventricles of the brain much distended with clear fluid, which spurted out when the ventricles were punctured, and was evidently under considerable tension. There was no evidence of thrombosis or any disease of vessels. No meningitis or thickening of the membranes discovered; no adhesions of pia-arachnoid to the dura mater; no disease of the bones of the skull or upper cervical vertebræ. The weight of the brain was excessive (1,588 grammes [1]—*i.e.*, 56 ounces—after being some time in formalin), but by macroscopic examination it showed nothing abnormal except an apparent thickening of the ependyma, which was most noticeable in the 4th ventricle. The choroid plexuses seemed healthy. The spinal cord was not examined. The stomach was not dilated, and there was no gastric ulcer or pyloric stenosis. Nothing noteworthy was discovered in the liver, kidneys, and the rest of the body.

Microscopic examination of the brain.—The upper part of the left ascending frontal convolution, part of the base of the brain, including a portion of the iter a tertio ad quartum ventriculum, part of the floor of the 4th ventricle close to the iter, and part of the medulla oblongata were examined. Sections were stained by the Weigert-Pal, Marchi, and Nissl methods. Sections of the ependyma from one of the lateral ventricles, from the third ventricle, and from different parts of the fourth ventricle were likewise examined, as well as a section of the choroid plexus of one of the lateral ventricles. All these sections were stained with logwood. The central canal of the medulla oblongata was, as it so often is, filled up with cells. The convolution showed nothing abnormal, unless, as Dr. F. E. Batten thought, slight "spacing of the nerve elements." The chief change was found in the ependyma of the fourth ventricle. The ependyma was infiltrated with round cells (fig. 1), and there was marked perivascular cell infiltration in the sub-ependy-

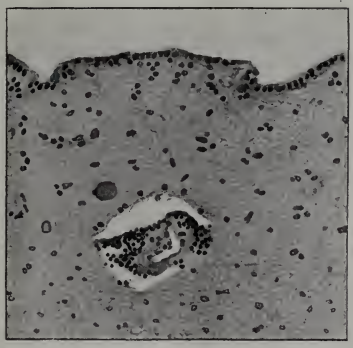
[1] This weight is decidedly excessive for a person of the patient's size, but it must not be considered very extraordinary. The brains of some notable men have weighed much more. Cuvier's is reported to have weighed almost 1,883 grammes (but see later footnote).



× 55 ½ Obj.

Fig. 1.

Section from floor of fourth ventricle, stained with hæmatoxylin. Ependymal surface rough and folded. No distinct epithelium at parts. Much cell-infiltration of the surface portion and around a sub-ependymal vessel (cut obliquely).



× 140 ¼ Obj.

Fig. 2.

A portion of the ependyma which is nearly normal (to compare with Fig. 1). Some perivascular cell-infiltration is however present—in the centre of the figure.

mal tissue. This inflammatory change was equally present near the iter and at the calamus scriptorius end, and was specially marked on the ependymal side of the membranous roof of the fourth ventricle and at the angle between the floor and roof. Sections from the ependyma of the third and lateral ventricles showed very much less change of this nature, the epithelium itself appearing nearly normal, though there was a little perivascular infiltration in the subjacent tissue. The section of the choroid plexus showed nothing very abnormal. A moderate number of the usual, more or less concentrically-marked, hyaloid or calcareous bodies could be seen, deeply stained with the hæmatoxylin ; and at one spot the epithelium of the capsule appeared thickened and infiltrated. The changes revealed by microscopic examination might therefore be almost summed up as *ependymitis of the fourth ventricle.* I must take this opportunity of thanking Dr. Batten for his help in the examination of the brain and microscopic specimens.

I have convinced myself that apparent thickening or slight granulation of the ependyma in cases of hydrocephalus should not, by itself, be accepted as evidence of a genuine inflammatory process. One must remember that the whole ependyma has been subjected to chronic stretching. The pressure exerted by the serous effusion must, of necessity, damage the tissues immediately bordering the ventricles, and will probably cause necrosis of individual cells. In chronic cases a reactive change will follow and will be accompanied by a certain amount of cell infiltration, as it is everywhere when a local necrosis occurs (*e.g.*, the changes around anæmic infarcts in the kidney[1]). Moreover, the macroscopic appearance of thickening, as well as the microscopic appearance of cell infiltration, will both be intensified by the collapse of membranous parts when the cerebro-spinal fluid is let out, and by the shrinking due to the action of hardening reagents.[2] In the present case, how-

[1] I think the cirrhotic changes in the liver resulting from obstruction of bile ducts by ligature or gall-stones can be likewise *partially* explained as a reaction to a great distension of the ducts.

[2] In the same way an apparent thickening and cell accumulation in the intima of blood-vessels may be produced by the hardening reagents employed in preparing the specimens for microscopic examination.

ever, there can be no doubt of the presence of true inflammatory changes in parts of the ependyma and sub-ependymal tissues. Moreover, the changes exactly resemble those figured by Burr and McCarthy as having been found in their case (to which I shall later on return), and as having been produced experimentally by the injection of irritating substances into the cerebral ventricles of cats.

The foregoing case is, in the main, similar to a case which I had the opportunity of seeing in Sir Dyce Duckworth's wards when acting as one of his medical clerks in 1887, and which he has kindly allowed me to refer to. The patient, a woman (A. S.) aged 57, suffered a considerable time, chiefly from severe headache, and had double optic neuritis. The necropsy ultimately showed the presence of simple internal hydrocephalus.

An apparently much more acute case than either of these was the following:—The patient, R. D., aged 21, a tailoress, born in Russia, was admitted on my side at the German Hospital in June, 1898. Eight days before admission she began to suffer from headache and attacks of vomiting. The headache was at first frontal and then occipital. She gradually became weaker, and just before admission had retention of urine. The general practitioner who had her sent to the hospital suspected a cerebral abscess. On admission, the pupils were found not to react to light and to be very small (pressure on the medulla?[1]) There was cervical rigidity. Temperature 98·8°F.; pulse 84, slightly irregular. Knee jerks natural. Urine, specific gravity 1020, no albumen or sugar. The patient died shortly after admission. At the necropsy the ventricles of the brain were found distended with clear fluid and the convolutions were flattened. There was slight thickening of the pia-arachnoid at the base of the brain, but nothing distinctly abnormal could be found. The lining of the lateral ventricles appeared slightly granular, but a microscopic examination showed nothing noteworthy. No disease was found in the heart, lungs, &c.

[1] Generally in adults with chronic hydrocephalus the tendency has been for the pupils to be dilated.

This case[1] may fairly be considered an example of acute internal hydrocephalus, similar to some of those described by Quincke and others, apparently resulting from serous meningitis.

Apart from what may be termed "passive hydrocephalus," "hydrocephalus ex vacuo,"[2] or "hydrops cerebralis ex vacuo," in which a portion or the whole of one or more of the ventricles is enlarged owing to some atrophic[3] condition of the neighbouring parts of the brain, cases of internal hydrocephalus may be divided into the following groups, the grouping being rather for convenience than for pathological correctness.

(1) Cases secondary to and a part of the phenomena of tuberculous[4] or any suppurative meningitis.

(2) Cases resulting from the presence of tumours, parasites, caseous nodules, abscesses, inflammatory foci, &c.[5]

(3) Traumatic cases.

(4) Cases of ordinary infantile or congenital hydrocephalus, whether or not associated with inflammatory thickening of the membranous roof of the fourth ventricle, involving Magendie's and the other foramina in it.

(5) Cases of internal hydrocephalus supervening on (or during convalescence from) epidemic or sporadic cerebrospinal (non-suppurative) meningitis. Of this group many typical examples have been recorded. Dr. E. P. Joslin,[6]

[1] Which I have already described in a paper in *St. Bart. Hosp. Reports*, vol. 34, p. 307.

[2] This so-called *hydrocephalus ex vacuo* may certainly be internal, and therefore is not merely synonymous with *hydrocephalus externus*, as Osler ("Principles and Practice of Medicine," 1901, fourth edition, p. 1028) gives it.

[3] The atrophy may be due to various causes. In certain cases there may at one time have been active internal hydrocephalus, and the pressure of the intra-ventricular effusion may have subsequently subsided.

[4] In some standard text-books and dictionaries the term "Acute hydrocephalus" is still given as synonymous with tuberculous meningitis, but this is altogether unjustified in the light of modern data on the subject.

[5] This class would include certain cases labelled "Meningitis serosa acuta," in which the exudation has not yet had time to become purulent, secondary to ear disease, &c. Compare Dr. J. Hegener, "Beitrag zur Lehre von der Meningitis serosa acuta," *Muenchener Med. Wochenschr*, 1901, No. 16, p. 617.

[6] *American Journal Medical Sciences*, October, 1900, pp. 444-463.

who gives an excellent account of the literature of the subject, says internal hydrocephalus has been known to be a result of epidemic cerebro-spinal meningitis since 1805, the year in which the disease is usually considered first to have appeared.

(6) Cases of so-called simple or idiopathic internal hydrocephalus of adults and older children due to a non-suppurative (serous) ependymitis or ventricular meningitis. See H. Quincke,[1] Gowers,[2] A. Heidenhain,[3] &c., on this subject.

A typical example of the so-called ventricular meningitis would seem to be the case described by Drs. C. W. Burr and D. J. McCarthy.[4] The patient, a male, aged 33, was suddenly seized with fever, cervical rigidity, headache, stupor and delirium. The fever fluctuated between 101° and 103.4° F. After three weeks, during which the intensity of the symptoms varied greatly, he improved much physically, but he was weak and his mental state was peculiar; he was altogether unlike his usual self, seeming rather like a man in the beginning stage of general paralysis. A week later the fever and meningeal symptoms returned, lasting about a week. Then after four days' interval they returned again and ended in death. *Post-mortem* examination revealed only a moderate internal hydrocephalus, proliferation of the ependyma and ependymal glia, perivascular round-cell infiltration in the sub-ependymal tissues, and sclerotic and degenerative changes in the choroid plexus. The authors give good plates of the changes observed and compare them to the changes produced by the injection of hydrochloric acid (five per cent.) or of sterile urine into the cerebral ventricles of cats.

My case (B. S.) belongs doubtless to this group, and the changes seen in the microscopic sections of the floor of the fourth ventricle correspond exactly to the changes in the ependyma and subependymal tissues described and illustrated by Burr and McCarthy in their case.

[1] Publications referred to later on.
[2] "Diseases of the Nervous System," 1893, vol. 2, p. 584.
[3] "Hydrocephalus acutus acquisitus internus," *Berliner med. Wochenschr.*, 1899, No. 49, p. 1078.
[4] *Journal of Experimental Medicine*, 1900, vol. 5, p. 195.

The whole subject of hydrocephalus and effusions into the ventricles can, I think, best be understood by comparison with effusions into the pleura or peritoneum. My first group corresponds to pleuritic effusions due to tuberculosis, or septic infection of the pleura. The second group is analogous to pleuritic effusions accompanying tumours, &c., close to or involving the pleura. The third group, traumatic hydrocephalus, seems hardly analogous to the results of injury to the thorax, though some cases may probably be compared to traumatic pleuritic effusions, containing blood mixed with serous fluid, following injury and bruising of the chest walls and lungs.[1] Chronic alterations in the walls of the cerebral vessels, due to chronic alcoholism, renal disease, &c., would doubtless sometimes play a part in the production of such traumatic cases. Perhaps, however, trauma should merely be considered as a possible exciting cause of hydrocephalus in children and adults, rather than as a basis for making a special group of cases ("traumatic"). Whatever the connection between trauma and hydrocephalus may generally be, there can be no doubt that a blow on the head has been the starting point of the symptoms in several well-recorded cases.

I shall now confine myself to the remaining three groups (4 to 6), and when speaking of internal hydrocephalus during the rest of the paper, my remarks should be understood as referring to these groups only. All the cases (excepting perhaps certain cases of infantile and congenital hydrocephalus, in which no evidence of previous posterior basal meningitis is found at the necropsy [2]) may be compared, I think, to non-suppurative pleuritis, that is, to pleuritis with serous effusion. The cases supervening on epidemic cerebrospinal meningitis (group 5) only differ from groups 4 and 6 in their connection with an epidemic disease, and therefore in their direct or indirect relation to the specific microbic

[1] Dr. J. Fawcett (*Trans. Path. Soc.*, London, 1897, vol. xlviii., p. 6) records the case of a boy aged 12, and gives other evidence in support of the view that a blow on the head may cause laceration of the iter, and lead to its cicatricial stenosis and to internal hydrocephalus.

[2] *Cf.* Lees and Barlow, in Allbutt's "System of Medicine," vol. vii., p. 545.

cause of the epidemic; but some cases of groups 4 and 6 are perhaps really cases following "sporadic" cerebro-spinal meningitis.[1] Between groups 4 (at least the cases showing signs of past meningitis) and 6 there is no clear pathogenic difference. Infancy is the favourite period for hydrocephalus to commence, and therefore far more cases fall into group 4 than into group 6, and, owing to the easy yielding of the skull[2] and consequent enlargement of the head, the diagnosis is more readily made in early life. Moreover, as will afterwards be shown, many cases classed with group 6, may quite probably be instances of acute exacerbations of chronic (more or less latent) hydrocephalus or else recurrences of the disease in apparently cured patients.

Professor Heinrich Quincke[3] has recorded a number of cases of hydrocephalus in adults, both fatal cases in which a necropsy was made and cases in which recovery took place, but in which the diagnosis was made more or less probable by the symptoms observed. By his description of cases and his work on spinal puncture he has doubtless contributed more than anyone else towards elucidation of the subject. He has grouped all cases of cortical or ventricular meningitis with serous effusion under the heading "serous meningitis" (meningitis serosa), and, as this name implies, he originally considered the serous effusion to be inflammatory. More recently,[4] however, he has laid stress on the possibly angioneurotic origin of *some*

[1] For instance, in H. Oppenheim's case (*Charité-Annalen*, Berlin, 1890, Jahrgang xv., p. 307) of a female, who died at the age of about 27, from an exacerbation of internal hydrocephalus, the first symptoms of which dated from the eighteenth year, the necropsy showed distension of all the ventricles and thickening of the ependyma and of the pia mater under the third ventricle, whilst the pia mater of a considerable area of the spinal cord was slightly adherent to the dura mater. So also in Eichhorst's case of a student, aged 23 (referred to in a later footnote), the membranes over the infundibulum of the brain were thickened, and the pia mater in the dorsal region of the cord was adherent to the dura mater.

[2] The head has, however, been known to increase in size from intracranial pressure after the time for closure of the sutures.

[3] See especially "Ueber Meningitis Serosa," in Volkmann's *Sammlung klinischer Vorträge*, Neue Folge, Innere Medicin, No. 23, 1893.

[4] See especially "Ueber Meningitis serosa und verwandte Zustände," by H. Quincke, *Deutsche Zeitschrift für Nervenheilkunde*, 1897, vol. ix., pp. 149-168.

cases, and Dr. W. Murrell [1] has likewise described a case which he believes to have been of angioneurotic nature.

This theory of an "angioneurotic hydrocephalus" seems to me, however—at least at present—to lack evidence to support it; and Quincke himself would probably only apply the angioneurotic theory to cases commencing suddenly without fever and of relatively short duration; analogous, in fact, to cases of angioneurotic œdema of the skin, and in which, perhaps, there might be an œdematous infiltration of the brain substance as well as an effusion into the ventricles.

Relatively high pressure of the intraventricular effusion is perhaps as consistent with the angioneurotic as with the inflammatory theory, but when the effusion of cerebro-spinal fluid is merely of angioneurotic origin, the specific gravity of the effused fluid and its richness in albumen should probably not be excessive. Following are a few records of the specific gravity of the cerebro-spinal fluid in cases of internal hydrocephalus. In a case of Quincke,[2] referred to in greater detail later on, the specific gravity of the fluid obtained by lumbar puncture was as high as 1015, and it contained a considerable amount of albumen. In a case related by Joslin [3] the fluid obtained at the necropsy showed a specific gravity of 1010, and contained one half per mille albumen. In a boy,[4] aged $8\frac{3}{4}$, apparently suffering from chronic hydrocephalus, the clear cerebro-spinal fluid obtained by lumbar puncture was of specific gravity 1008, and contained one half per mille albumen. In a boy[5] 9 years old, lumbar puncture gave a clear fluid of specific gravity 1008, containing three-quarters per mille albumen. In two hydrocephalic infants[6] the cerebro-spinal fluid obtained by

[1] *Lancet*, April 28, 1900, p. 1,206. Murrell uses the term, "Quincke's Disease," but surely, if used at all, this term should signify "Angioneurotic œdema of the skin," in connection with which Quincke's name is so well known.

[2] *Verhandlungen des X. Congresses für inn. Med.*, Wiesbaden, 1891, p. 333.

[3] *Loc. cit.*, p. 449, Case iii.

[4] Quincke, Volkmann's *Sammlung*, *loc. cit.*, p. 664, Case 9.

[5] Quincke, Volkmann's *Sammlung*, *loc. cit.*, p. 666, Case 12.

[6] Quincke, Volkmann's *Sammlung*, *loc. cit.*, pp. 687-9, Cases 1 and 2. In the first case the fluid was obtained by tapping a spina bifida in the lumbar region, but at the necropsy the fluid distending the lateral ventricles was found shut off by occlusion of the iter. Recently I had the opportunity of examining the fluid obtained by tapping the lateral ventricle of an infant with ordinary chronic hydrocephalus. It was clear, of specific gravity 1005, giving a thin cloud of albumen and slightly reducing Fehling's solution.

lumbar puncture was, in the first case (from a spina bifida), of specific gravity 1007, with half per mille albumen; in the second case, of specific gravity 1009, with three-quarters per mille albumen. In seven other patients referred to by Quincke,[1] between 12 and 41 years of age, the cerebro-spinal fluid obtained by lumbar puncture had a specific gravity of 1007-1010, and contained one half to 2·7 per mille albumen.

On the whole, it is best to consider the angioneurotic theory of hydrocephalus as not well supported, and it is questionable whether the theory is required to account for any of the cases. For it must be remembered that only a limited portion of the ependyma and meninges need be inflamed to cause an effusion. The macroscopic appearance of thickening of the ependyma is not necessarily due to cell infiltration, and the presence or absence of this infiltration cannot be determined without microscopic examination. Thus, in my present case (B. S.) a portion of ependyma, examined microscopically, showed practical absence of cell infiltration, though it appeared thickened like the infiltrated portion. When there is a more or less general appearance of thickening or granulation of the ependyma, owing to shrinking after relief of the tension (due to escape of the cerebro-spinal fluid at the necropsy), the evidence of real inflammation might be overlooked, owing to a portion of the ependyma being selected for microscopic examination which happened not to show typical cell-infiltration. In Prof. H. Eichhort's [2] case of a student, aged 23, who, after an illness of a few months' duration, died with symptoms of cerebral compression, which the necropsy showed to be due to internal hydrocephalus, it is expressly stated that the ependyma was not universally much thickened, but in numerous small patches only. In some cases of localised ependymitis the effused fluid might even not be of particularly high specific gravity or particularly rich in albumen. For the fluid effused from the limited inflamed surface might have an irritating quality, leading to the copious pouring-out of normal, or nearly normal, cerebro-spinal fluid from the

[1] *Deut. Zeit. f. Nervenheilkunde, loc. cit.*, Cases 1, 2, 3, 4, 6, 8, 9.
[2] *Zeitschrift für klin. Medicin*, Berlin, 1891, vol. xix., Supplement, p. 181.

non-inflamed surfaces.[1] Moreover, local conditions of the circulation (venous and lymphatic obstruction, by pressure or otherwise, in the choroid plexuses and venæ Galeni) would doubtless influence the rapidity and pressure of the effusion and the quality of the fluid effused. Thus sudden changes in position of the patient's body, alcoholic stimulants, exposure to heat or cold, mental or physical exertion, would all exercise an influence. To the probable effect, in this respect, of closure of Magendie's foramen, I shall refer later on. General conditions, such as renal disease and chronic alcoholism, producing alterations in the blood and blood-vessels, would likewise, if present, have an influence on the effusion.

The difficulties of the whole subject can best be exemplified by returning to the comparison with serous effusions into the pleura and peritoneum. Chance necropsies after attacks of pleuritis show that only limited portions of the pleura need be much affected. In fact, in serous pleuritis the inflammation may be very unequally distributed over the serous membrane, and a small portion only of the pleura need be inflamed to give rise to a serous effusion of the whole pleura. These circumstances by themselves, independently of important local and general conditions of the circulation, may account for much of the variation in the quality of the effused fluid observed in cases of serous pleuritis.

Like many others, I think that ascites occurring in cases of early hepatic cirrhosis and cardiac disease is frequently entirely or partially due to inflammatory changes; but I believe that the reason why this has not been generally admitted is that the peritoneal inflammation in such cases (a serous peritonitis analogous to a serous pleuritis) is often limited to a very small area[2] only of the peritoneal surface, though it gives rise to a general ascitic effusion. If this be

[1] From a "teleologic" point of view, the fluid from the non-inflamed surfaces is poured out with the object of diluting the irritating fluid poured out from the inflamed surface.

[2] For instance, there may be only a little perihepatitis or perisplenitis, as I remarked at the discussion of Dr. Campbell Thomson's paper at the Royal Medical and Chirurgical Society (*Lancet*, June 15, 1901, p. 1686).

recognised and the influence of abnormal conditions of the general and local circulation be likewise considered, one cannot wonder that the specific gravity of the ascitic fluid in such cases may vary considerably, that it may be in some cases as high as 1016 to 1020, though in other cases it is only 1010 or 1012 or thereabouts. Yet I believe the specific gravity of the effused fluid may sometimes be helpful in recognising the inflammatory origin of such effusions, whether in the peritoneum, pleura or cavities of the central nervous system.

I come now to the question of the inflammatory closure of the foramen of Magendie and the neighbouring foramina in the roof of the fourth ventricle between the cerebellum and medulla, and to the possible influence of this closure (or of inflammatory thickening and consequently lessened permeability of the whole membranous roof of the fourth ventricle) in the pathogeny of internal hydrocephalus. *Post-mortem* observations show that thickening of this region is not invariably present in cases of internal hydrocephalus, and therefore the change in question cannot be regarded as the only essential factor in the pathogeny of the hydrocephalus. The reason why inflammatory thickening or "plastering" of the membranous roof of the fourth ventricle is almost invariably found *post mortem* in cases of internal hydrocephalus supervening on epidemic or sporadic cerebro-spinal meningitis, such as those described by Joslin,[1] is probably that the region is one of the sites of election in the kind of meningitis in question. It can scarcely be doubted, however, that the inflammatory thickening of this region, when present in any case, may be regarded as a factor in the production of the hydrocephalus, though of course it can have no influence in the rare cases in which the fourth ventricle is not involved in the hydrocephalic distention and in which there is obstruction in the iter[2] or closure of one of the lateral ventricles.[3]

[1] *Loc. cit.*

[2] Blocking of the iter, doubtless sometimes congenital, may be due to tumours or parasites or inflammatory changes, or apparently to cicatricial stenosis resulting from traumatism, as in Dr. J. Fawcett's case (*loc. cit.*).

[3] E. Ziegler, *Lehrbuch der spec. Path. Anatomie.* Ninth edition, 1898, p. 332.

I have now to consider certain points of interest connected with internal hydrocephalus in adults and elder children, the first being the fact that, as Quincke has pointed out, apparently acute "idiopathic" cases are in reality sometimes only exacerbations of a chronic condition, which occasionally even dates from early childhood or infancy. As examples I will quote a few cases, and first of all the well known one described by John Hilton.[1]

A gentleman, who died at the age of 34. As a child he was active, but irritable in temper; as a man he was very spare and delicate looking. The terminal phalanges of his fingers were very short, and the nails inserted like seeds into the cuticle. He was fond of intellectual pursuits, and had a great dislike to excitement and noise. Excitement frequently brought on an impediment in his speech. At the age of 16 he had a severe nervous illness with depression, said to have been brought on by application to business in the City, which, however, was not of a kind to affect ordinary persons. Tea, coffee, and cocoa seemed to affect his head and derange his stomach, and he had an instinctive dislike to alcoholic stimulants of every kind. As he said himself, he only half lived in the winter; he seemed torpid, and would drop into a deep sleep after a meal, from which it was very difficult to rouse him at bedtime, when he seemed scarcely to know where he was. Though he was subject to coldness of the extremities, and though both winter and cold always affected him injuriously, yet he could not bear a warm room, as it made him feel faint. For many years he was subject to headache, derangement of stomach, and occasional deafness. His pulse was usually weak, irregular, and somewhat infrequent, about 50 to 60 beats in the minute. A peculiar uneasy look of the eyes, with dilated pupils and muddiness of the conjunctiva, was said to be noticeable when his health was temporarily deranged. During the last year of his life he gradually lost flesh and became more feeble, acquiring a slight stoop in his gait. He also seemed to be affected with slight stiffness of

[1] "Influence of Mechanical and Physiological Rest." First edition, London, 1863, p. 39. Some points in this history suggest that the patient may have had syringomyelia as well as hydrocephalus.

the neck. A few months before his death he had a severe attack of vomiting, with prostration, without obvious cause. On the day of his death he had been out, about a mile or so, from home, and had twice been seized with vomiting. He walked home, but felt giddy and oppressed, and was placed on a bed. He died in a short time with stertorous breathing, although he remained sensible almost to the last moment. At the *necropsy* the cerebral convolutions were found flattened, and were rather large and few for the man's age, suggesting a certain degree of arrested development. The lateral ventricles contained at least four ounces of clear cerebro-spinal fluid. The fourth ventricle was greatly dilated in all its directions. The septum lucidum and fornix were softened, but the microscope showed no trace of inflammatory deposit. There was a tolerably dense membranous structure between the under surface of the cerebellum and the upper surface of the medulla oblongata, closing the cerebro-spinal aperture and forming a kind of pouch projecting downwards, thus showing the direction of the fluid tension upon it to have been from above downwards. Hilton concluded that the gastric trouble and occasional deafness were due to distension of the fourth ventricle, with consequent pressure on the origins of the pneumogastric and auditory nerves.[1] "His brain could bear no fulness of blood, no increase of size, because the cerebro-spinal fluid could not escape from the interior of the brain. He could not take wine, beer, or spirits;[2] and could bear no muscular

[1] In Burr and McCarthy's case, already alluded to, deafness was one of the symptoms, and, *post mortem*, the eighth pair of cranial nerves were found very much degenerated.

[2] Compare this case with the case recorded by F. Plehn (quoted by H. Quincke, Volkmann's *Sammlung*, *loc. cit.*, p. 670). A student ever since his tenth year, when he was rendered unconscious by a fall, suffered every three to eight days from attacks of severe headache, lasting several hours. Speaking or the application of cold to the head during the attacks made them worse, and he was very careful in regard to alcohol, owing to its increasing his headaches. At the age of 23, after spending an evening with his friends, he was seized with a worse headache than usual, followed by loss of consciousness, Cheyne-Stokes breathing, and death. The necropsy showed distension and dilatation of all four cerebral ventricles, and thickened ependyma. There was a sacculated dilatation of the fourth ventricle at the point of the calamus scriptorious. The pia mater was thickened over the posterior part of the cerebellum, and the foramen of Magendie seemed to be obliterated. Fagge see Fagge and Pye-Smith, "Medicine," 1901, vol. i., p. 883) recorded the case of a bank porter whose symptoms had commenced seven years previously, after a blow on the head. Ever since the injury he had been liable to attacks of occipital pain and vertigo; reading often made him feel giddy, and he could scarcely take any stimulant.

exertion. The congestion of the brain produced vomiting by pressure upon the medulla oblongata, exerting its influence through the pneumogastric nerve."

The next case I have selected is one recorded by Professor H. Quincke[1] at the German Medical Congress at Wiesbaden, in 1891.

The patient was a man, aged 25, who had often suffered from headache since December, 1889, and from vertigo since the spring of 1890; in the following June optic neuritis was noted. There was no history of previous illness, but the least indulgence in alcohol or tobacco had always made him feel giddy. On admission into the hospital, November 7, 1890, the patient, a strongly-built, big man, suffered from headache and vertigo. Both eyelids were moderately swollen. Optic neuritis. No paralysis of limbs. The diagnosis rested between cerebral tumour, chronic meningitis, and exacerbation of an old hydrocephalus. Ice-bags, purgatives, iodide of potassium and mercury had no thorough effect, though the headache almost went away, and at times the vertigo was much diminished. Counter-irritation gave some relief. On April 3, 1891, the cerebro-spinal fluid was tapped by lumbar puncture, the manometer pressure being 50 cm. water, equal to 37 mm. mercury. In the course of an hour 80 cc. of clear fluid came away, of specific gravity 1015, containing a considerable amount of albumen. On the following day there was less vertigo.

Quincke regards the case as an exacerbation of a hitherto latent chronic hydrocephalus, and it seems to me that the character of the cerebro-spinal fluid removed by spinal puncture throws light upon the nature of the exacerbations in such cases.

The following case, recorded by Dr. Frederick Taylor,[2] is instructive, because the patient, who died from an exacerbation of chronic hydrocephalus at the age of 16, seemed to show signs of congenital hydrocephalus by the large size of

[1] *Verhandlung des X. Congresses für innere Medicin*, Wiesbaden, 1891, p. 333. This case may be compared to some of those described by Quincke in Volkmann's *Sammlung*, *loc. cit.*, and in the *Deut. Zeit. für Nervenheilkunde*, *loc. cit.*

[2] *Transactions of the Clinical Society of London*, 1897, vol. xxx., p. 175.

his head at birth. It is unfortunate that the weight of the brain at death is neither mentioned in this nor in Hilton's case; both the patients in question gave evidence of considerable intellectual development, and possibly had slightly "hypertrophied" brains, such as those alluded to later on.

Dr. Taylor's patient was a boy aged 16, admitted into Guy's Hospital, September 11, 1895. His head was large from birth. At 8 months he had bronchitis and fits. He did not walk properly till he was 1 year and 10 months old. He talked plainly at $2\frac{1}{2}$. At 8 he had scarlatina, which was followed by nocturnal incontinence of urine. At 12 he learned the violin, and could play it and sing well. At $13\frac{1}{2}$ he left school, and became a compositor, working regularly about 54 hours a week. His mother considered him to be strong and active, and quick at picking up information. Headache and vomiting were the prominent symptoms in the hospital. Headache had been first complained of in August, 1895, and vomiting on September 2. His gait was unsteady. There was no anæsthesia or loss of sensibility to pain or temperature. Knee-jerks somewhat increased on both sides. Optic discs normal. The headache was constant across the forehead, and occasional over the occiput, sometimes very severe, especially on sitting up. On September 16 the head was retracted, and during the next days patient was drowsy. On September 19 there was severe pain in the back of the neck. Weakness in legs. On September 20 there was a little fever. Weakness in all limbs; increasing in the legs. Knee-jerks absent. Slightly drowsy. Speech hesitating. Some nystagmus. No optic neuritis. Embarrassed breathing. Cyanosis. Death. At the necropsy the bones of the skull were found to be remarkably thinned by pressure. The pia arachnoid was thickened over the base of the cerebellum, particularly near the foramen of Magendie. It was adherent to the dura mater in several places, especially over the foramen magnum. There were thirty ounces of liquid in the distended ventricles. The ependyma was granular.

Occasionally hydrocephalic patients, with heads already greatly enlarged in childhood, have lived to adult life. A

well-known example of this is the man James Cardinal, the subject of chronic hydrocephalus from infancy, who died at Guy's Hospital in 1825 at the age of nearly 30 years, probably of some different disease.[1] It is very probable that some of the persons who have been celebrated on account of the excessively large size of their heads are analogous examples. M. A. Ruffer[2] refers to cases in which a considerably greater age than that of Cardinal was attained. Dr. W. A. Brailey's case[3] was a woman who died of acute pneumonia at the age of 53; and Trousseau[4] mentions two old men, aged 72 and 78 respectively, whose history was known to Frank, and who were hydrocephalic from birth.

Though in my present case (B. S.) there is no history pointing to the early presence of hydrocephalus, as there is in some of the cases I have just quoted, yet the overgrowth of the brain, shown by its weight, points to the fatal illness being probably a complication of a much older disease. It brings the case into connection with those of the so called "hypertrophy of the brain," or "megalencephaly," an excellent example of which was shown in 1900 to the Pathological Society of London by Dr. H. Morley Fletcher.[5] L. Edinger[6] says that his friend Perls first conjectured that the pressure exerted on the brain by the skull might be reduced in cases of hydrocephalus which are undergoing cure, and that the brain substance might, as a result of not being opposed by the normal pressure of the skull, increase abnormally in bulk.

One may suppose that if this conjecture be correct, and if such increase be due merely to overgrowth of the connective tissue elements, it may lead to mental backwardness

[1] Possibly of pulmonary and intestinal tuberculosis. See the account given of Cardinal's case by Dr. Richard Bright, *Reports of Medical Cases*, London, 1831, vol. ii., p. 431.

[2] "Critical Digest on Chronic Hydrocephalus," BRAIN, 1890, vol. xiii., p. 251.

[3] *Trans. Path. Soc. of London*, 1881, vol. xxxii., p. 1.

[4] "Clinical Medicine," New Sydenham Society's Translation, London, 1868, vol. i., p. 475.

[5] "A case of Megalencephaly," *Trans. Path. Soc., London*, 1900, vol. 51, p. 230.

[6] "The Anatomy of the Central Nervous System," English translation, by Dr. W. S. Hall, 1899, p. 206.

or defective intelligence and to imbeciles with heavy brains; but if the nobler elements participate equally in the excessive growth unusual intellectual powers may, as Perls and Edinger have suggested, be the result. In my case (B. S.) there may have been a hydrocephalic tendency in early life, the brain afterwards undergoing abnormal increase in size[1] as a consequence of recovery. The later disease may have been a recurrence started by the second attack of erysipelas.

There are cases of internal hydrocephalus in elder children or adults in which *post-mortem* examination has demonstrated that a condition of hydromyelia or syringomyelia of the spinal cord has likewise been present. Dr. Pierre Marie[2] recorded the case of a man, who died at the age of 36, in whom at the *post-mortem* examination a condition of hydrocephalus was found associated with extensive hydromyelia or syringomyelia of the cord. I have likewise heard of two cases of fatal hydrocephalus, in which at the necropsy the presence of syringomyelia or hydromyelia in some part of the spinal cord was ascertained. In the same way hydrocephalus in infants has long been[3] recognised as not rarely associated with spina bifida, a congenital abnormality certainly to some extent allied to hydromyelia and syringomyelia, even though the last mentioned condition can hardly be termed congenital.[4]

The foregoing considerations all lead one to suspect that some of the cases described as "idiopathic" acute internal hydrocephalus in adults (the *post-mortem* examinations being often only partial) may in reality have been exacerbations in chronic cases, or at least connected with old abnormal conditions of the central nervous system.

I now come to another matter, namely, the rare occurrence of symptoms of internal hydrocephalus in persons

[1] It is unfortunate that we have not more records of the weights of hydrocephalic brains from patients of various ages. Hervé suggested that Cuvier was affected with hydrocephalus in his childhood. At the *post-mortem* examination it was found that the lateral ventricles were very large. According to E. Rousseau's account the weight of his brain was 1861·2 grammes, but according to Bérard's official report it was still greater, 1882·9 grammes. (See *Revue de Thérapeutique*, Paris, December, 1883, page 623.)

[2] Société de Neurologie, Paris, December 7, 1899.

[3] See Dr. Richard Bright, *op. cit.*, vol. 2, p. 428.

[4] See however H. Schlesinger's classification (*Syringomyelia*, 1902).

with a myxœdema-like puffiness or œdema of the face. When a student, in 1887, I saw a young man, aged 18, who died at the German Hospital with internal hydrocephalus, the *post-mortem* examination showing no other cerebral disease except distension of the ventricles with cerebro-spinal fluid. The puffiness of the skin in his case was so remarkable that the diagnosis of myxœdema had been made by a great authority on the subject. In Quincke's case, of which I have already given an abstract, the eyelids are stated to have appeared swollen. I should have regarded the myxœdema-like condition in the young man as a coincidence, were it not that I afterwards read Quincke's paper, and that in November, 1900, my colleague, Dr. zum Busch, kindly showed me a small child, under his care at the German Hospital, in whom a striking myxœdema-like puffiness of the face was noted, together with symptoms of cerebral compression, supposed to have followed an injury. The cerebral symptoms improved after spinal puncture. Whatever the explanation of these facts may be, it must be admitted that the occasional facial œdema and the exciting influence of traumatism lends some, though I think insufficient, support to the theory of the existence of an angio-neurotic hydrocephalus. In this connection I may likewise draw attention to certain cases in which a chronic condition of solid œdema about the eyelids and face is associated with recurrent erysipelas-like attacks. My patient (B. S.) was stated to have suffered previously from erysipelas; and in a girl, aged 14, whose case is described by Quincke,[1] the cerebral symptoms commenced after an attack of facial erysipelas, though the relatively large head of the patient suggested the possibility of previous latent chronic hydrocephalus.

The *chief symptoms* of hydrocephalus in adults appear to be: headache, stiffness in the neck, sudden violent attacks of vomiting, double optic neuritis, weakness, and a hesitating gait. In the present case the changes shown by ophthalmoscopic examination were not very great, but in the case of a

[1] Volkmann's *Sammlung*, *loc. cit.*, p. 659, Case 4.

labourer, aged 31, recorded by Annuske,[1] in which death occurred after an illness of about five months, and in which the necropsy showed great dilatation of all the ventricles, blindness was complete in about three months. Some of the cases which recover show quite as marked ophthalmoscopic changes as fatal cases do; for instance, in a woman, aged 22, recovery from the brain symptoms took place after ten months, but was accompanied by amaurosis due to optic nerve atrophy.[2] In some of the necropsies the floor of the third ventricle and the infundibulum have been found much bulged out by the hydrocephalic effusion, showing the special pressure to which the optic tracts and the optic chiasma must have been subjected. In Taylor's case, however, to which I have already referred, there was no optic neuritis, even shortly before the patient's death. Other cranial nerves beside the optic are sometimes affected. Thus, in Hilton's case and that of Burr and McCarthy, both already referred to, the auditory nerves suffered; in a young woman, the subject of a paper by Oppenheim,[3] there was complete anosmia, and at the necropsy the olfactory nerves were found much flattened out.

The vomiting was sometimes very violent in the present case. A notable feature was the tendency to vomit on lying down after being in the upright position. This effect of lying down suddenly may be accounted for if we remember that the change of position favours the flow of blood to the head, and thus adds to the pressure, due to the hydrocephalic effusion, to which the brain is already subjected. Some of the symptoms noted in Hilton's case may be compared to it. Oppenheim on one occasion, in the case to which I have just referred, noted that the patient continually held her head forwards and to the left, because attempting to move it to the contrary position immediately induced vomiting.

[1] von Graafe's *Arch. f. Ophthalmologie*, Berlin, 1873, vol. xix., Part III. p. 260, Case 6. It is quoted by Quincke, Volkmann's *Sammlung*, *loc. cit.*, p. 669.

[2] Quincke, *Deut. Zeit. f. Nervenheilkunde*, *loc. cit.*, p. 150, Case 2.

[3] The case (*Charité-Annalen*, *loc. cit.*) has already been quoted in a footnote.

Before the onset of the cervical rigidity, the vomiting in the patient, B. S., was supposed to be due to gastric disease. Fagge,[1] amongst various cases of adult hydrocephalus at Guy's Hospital, mentions that of a man aged 55, who had complained of severe pains in the head three months before his death, and was admitted into the hospital for symptoms resembling those of chronic dilatation of the stomach. He died in a semi-comatose condition a few hours after a convulsion. Strauss[2] mentions the case of a man aged 50, who died with symptoms resembling those of cancer of the œsophagus. At the necropsy nothing could be found to account for the œsophageal obstruction, but chronic internal hydrocephalus was present. During life, however, there had certainly been obstruction near the cardia to the passage of the œsophageal bougie, and it must, therefore, have been due to œsophageal spasm of nervous origin, probably connected with the condition of hydrocephalus. In my case (B. S.) the reported absence of free hydrochloric acid from the gastric contents after test meals may possibly be accounted for as of nervous origin, and connected with the hydrocephalus. The condition of "achylia gastrica," though certainly usually associated with organic disease of the stomach, may probably sometimes develop, as writers on the subject have pointed out,[3] from nervous disturbance.

The terminal convulsive seizure in my case, as in others, was probably due to sudden increase of intraventricular pressure acting through pressure on the floor of the fourth ventricle.

In regard to *treatment*, the increase in the morbid symptoms or relapses which have sometimes been observed to follow muscular over-exertion or other kinds of over-fatigue, or even small doses of alcohol, furnishes us with some useful hints for caution, especially in regard to improving cases. There cannot be a doubt that great improvement and even apparent recovery from the symptoms

[1] Fagge and Pye-Smith's "Medicine," 1901, vol. i., p. 832, Case 4.
[2] *Berliner klin. Woch.*, September 19, 1898.
[3] See Max Einhorn, "Diseases of the Stomach," Second Edition, 1898, p. 328.

of hydrocephalus in adults occasionally takes place, with or without any special treatment beyond rest in bed, &c. In the present case the improvement was at one time so great that the question of the patient being able to leave the hospital was beginning to be considered. Joslin[1] is not convinced of the usefulness of lumbar puncture in hydrocephalus (at least in his cases following meningitis), but Quincke's results seem to indicate that it may sometimes be of real use, apart from diagnostic purposes, by the temporary relief of pressure.[2] Mercury, iodide of potassium, and possibly salicylates and antipyrin, as well as counter-irritation, are worth a trial, and mercurial treatment epecially may be of service. The improvement sometimes following treatment by calomel, mercurial inunction, and iodide of potassium might in these cases, as in various other brain cases, occasionally lead to an erroneous diagnosis of syphilis.[3]

Following are some conclusions on the subject of hydrocephalus which I believe are justified :—

(1) That the various kinds of hydrocephalus and effusions into the ventricles of the brain may be fitly compared to the various kinds of effusions into the pleura and the peritoneum.

(2) That the cases of so-called idiopathic or simple internal hydrocephalus are probably nearly all due to more or less localised serous meningeal or ependymal inflammation and are strictly analogous to cases of serous effusion into the pleura or peritoneum, resulting from localised non-suppurative pleuritis or peritonitis. It is of course probable that there are several different microbic or toxic agents.

(3) That the reason why chronic inflammatory thickening of the membrane stretching from the cerebellum to the medulla and forming part of the roof of the fourth ventricle

[1] *Loc. cit.*

[2] Lumbar puncture for the relief of intraventricular pressure is probably of greatest service in the blood-stained serous effusions resulting from traumatism.

[3] Hydrocephalus in infants in a few cases has been undoubtedly connected with and caused by congenital syphilis. This has been insisted on especially by French writers, and improvement and even cure have been observed under antisyphilitic treatment. See Marfan "L'hydrocéphalie des nourrissons," *La Semaine Médicale*, Paris, 1898, p. 193.

has so often been found present in fatal cases of chronic hydrocephalus, is not necessarily merely that the foramen of Magendie and the neighbouring foramina have been closed by inflammation (as they very often must be in similar cases), but that this portion of the roof of the fourth ventricle is one of the sites of election for the localised inflammation which leads to hydrocephalus.

(4) That the theory of a purely angioneurotic effusion to account for some cases of acute internal hydrocephalus has as yet not sufficient evidence to support it, though doubtless the amount of irritation required to produce the same pressure of effusion varies much in different individuals; it varies doubtless according to conditions (temporary or persistent, congenital or acquired) of the blood-vessels and lymphatics which influence the local circulation; doubtless also temporary circumstances, such as sudden change from the upright to the recumbent position and *vice versâ*, menta or physical exertion, undue exposure to cold or heat, the ingestion of alcohol or other stimulants, and reflex effects on the local circulation from other parts of the body, may influence the tendency to effusion in the ventricles.

(5) That many of the cases of apparently acute hydrocephalus in adults and older children are really exacerbations of a chronic condition, sometimes doubtless dating from early childhood or birth, as evidenced by the history of previous cerebral symptoms, by the relatively large size of the head, or by the *post-mortem* evidence of association with a condition of hydromyelia or syringomyelia, or, as in the present case, by excessive weight of the brain.

CONTENTS.

ORIGINAL ARTICLES AND CLINICAL CASES:—

REVIEWS:—

[ELSSON, LTD., PRINTERS.] [83-89, GT. TITCHFIELD STREET, LONDON, W.